EARLY RACHITIC CHANGES IN THE FEMUR AND TIBIA

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Although the etiology of rickets is still unknown, and its early symptoms are frequently very obscure, an early diagnosis may be definitely made by means of the roentgen rays, and treatment started before the development of structural deformities.

Full discussion by William Palmer Lucas, San Francisco; Rolla G. Karshner, Los Angeles; William Sidney Bowers, Los Angeles; H. H. Markel, San Francisco.

THE etiology of rickets—a disease which may exist without symptoms, but which early affects the structure of the bones—is not known, and it is, therefore, very difficult to recognize and study in its early stages. Of the various theories advanced, that of defective feeding seems to offer a cause in the greater number of cases, the diet being so unbalanced that it really seems antagonistic to the deposition of calcium in the growing bone matrix.

After a most thorough review of the literature, one is unable to select a diet that will prevent rickets in every person. A low-grade infection, lack of sunshine and fresh air, insufficient amount of exercise, prenatal influences, or some constitutional disorder, apparently causes rachitic changes in cases where the diet has been carefully selected. On the other hand, rickets is frequently found in a seemingly well child of healthy parents where the diet has always been well-balanced, fresh air and sunshine have been abundant, and there was a normal amount of exercise each day. Rickety children are often fat, rosy, and apparently well nourished.

In normal developing bone, the areas of ossification, and the calcification, proliferation and arrangement of the cartilage cells are definitely marked off from each other. This orderly arrangement is lost in rickety bone, as the lime salts are deficient in some parts of the cartilaginous matrix, and irregularly deposited in others. There is also a marked increase of the proliferating cartilage cell zone, which, together with an overproduction of the osteoblastic elements, results in enlargement of the bony structure, but of an imperfect character because of the deficiency of the lime salts.

The growing line at the epiphyseal junction is broader than normal, the width also being considerably increased and easily felt when the ossifying cartilages are subcutaneous. There is excessive proliferation of the osteoblastic layer of the periosteum, which increases the circumference of the bone. Under proper treatment the rachitic changes give way to a more normal process of ossification. When discovered and treated early, the rickety bone frequently shows but slight changes from normal bone.

The large epiphyses, bent limbs, large square head, Harrison's groove, rickety rosary, "pot-belly," delay in closure of the anterior fontanelle, profuse head-sweating, delayed dentition, and marked muscular weakness, give a clinical picture so striking that rickets can hardly be mistaken. However, in mild cases it is difficult to determine what constitutes evidence of rickets.

Occasionally there are no symptoms except bow-legs (genu-varum) or knock-knees (genu-valgum), which mother felt sure did not exist a few weeks ago. More frequently, vague symptoms with gradual onset, and general distribution, do not suggest rickets until there have been sufficient changes in the bones to cause the characteristic deformities. Abnormal lateral mobility and hyperextension of the knee joint, resulting from relaxation of the ligaments and from weakened condition of the muscles, contribute largely to bow-legs or knock-knees. The latter are also aggravated by the everted feet, so frequently seen in these cases.

Occasionally there has been no decrease in activity. More often there has been for some time vague symptoms which did not yield to treatment which, however, was not for rickets. An x-ray of the lower extremities apparently would have been the only definite way of determining the cause of the symptoms. It is this type of rickets so frequently overlooked, even by excellent diagnosticians, and responding quickly and rather completely to treatment, which I wish to emphasize in this paper.

An x-ray on one plate, from just below the crests of the ilia to and including the feet, is necessary, not only to determine the changes in the femur and tibia, but to give an accurate record of the deformity. In these very early cases the greater changes are at the lower end of the femur, and upper end of the tibia. The whole joint has a hazy appearance, the shafts near the epiphyseal lines are clearer and more transparent than normal. There is also a noticeable thickening at these points. The epiphyseal lines are considerably broader and not nearly so clearly defined as normally. Usually there is an increase in the calcified material in the epiphyseal region. The lower epiphysis of the tibia and fibula show changes similar to those just described, sometimes more marked, but frequently not nearly so noticeable.

Operative interference is not indicated in any type of rickets where the bones are still soft. Nor is it necessary to subject the patient to the prolonged use of braces. During the acute stage there is hyperemia of the entire joint, which should be protected for several weeks by complete rest.

The type of rickets which has just been described responds quickly to the following treatment. Very slight changes, if any, are made in the diet, but cod-liver oil is given for several weeks. Exposure to sunshine in the open is a routine.

Corrective treatment is begun immediately. By means of thick chamois-covered felt pads, placed between the internal condyles, in knock-knees, the internal malleoli are gradually brought together while the knees are held firmly to the table. This overcorrection is maintained about five minutes, then light plaster of paris casts are applied as rapidly as possible from the groin to and including the foot.

In this way correction is obtained with the first casts, without using any pressure during their application. No weight-bearing for four weeks. Then another stretching gives full overcorrection which is maintained for four weeks by walking-casts. The muscle balance is restored by high shoes with one-eighth of an inch float on the inner side of the heels and soles, and frequent stretching at the knees. In from two to four months after removal of the casts

correction is not only complete, but the x-ray gives evidence of normal or greater density of the bones.

A simple device, fastened to the table, keeps the knees fully extended during the stretching. This is made of two pieces of aluminum, twelve inches long, five inches wide, padded with felt and covered with chamois. The lower one is fastened to the table by two bolts with thumb screws, while the upper one is held firmly against the knees by two pieces of webbing passing through each end of the pads and buckling on top.

By gradually increasing the pads between the internal condyles, marked overcorrection may be obtained and held ten or fifteen minutes without discomfort to the patient, especially if the outer part of the ankles is protected by felt pads. As the appliance which holds the knees firmly to the table allows about 5 degrees of flexion, there is no unnecessary strain to the posterior part of the knee joint during the stretching.

Since the process is much more difficult in cases of bow-legs, the following appliance has been very helpful. Two small semi-circular pieces of metal, to one side of which is riveted a strap, then covered with felt, and to the other a narrow piece of metal four inches long with several screw holes, are fastened as closely together as possible. On the other end of each strap is a sliding felt pad, to complete the circumference of the leg. This appliance, which may be gradually widened to seven inches, is strapped on just above the ankles. The feet are held markedly everted by an assistant while the knees are slowly brought to a position of overcorrection and held there several minutes.

The procedure with casts is the same as with knock-knees, but the float on the shoes is on the outer side of the heels and soles. The feet must be carefully watched and the floats removed when the first sign of weakness appears.

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DISCUSSION

[EDITORIAL NOTE—*Ordinarily, discussions of papers are limited to five hundred words. However, an exception is made in the case of Doctor Lucas' discussion, because of the tremendous importance of the questions raised by Doctor Chappel, and because of the additional opinion brought to bear upon the question as the result of original investigation.*]

WILLIAM PALMER LUCAS, M. D. (University of California Medical School, San Francisco) — Doctor Chappel's discussion of the pathology of the bone in rickets is very well expressed. I am not in a position to discuss his corrective treatment and orthopedic stand, but concerning the etiology of the disease, although the cause is still obscure, I do not think we need to be as pessimistic as one would be led to believe from Doctor Chappel's introductory paragraph. Since Mellanby published his experimental work in 1915 on puppies, many very valuable contributions to our knowledge of rickets have been brought forward, and we are in possession at the present time of means of successfully preventing and treating early rickets before deformities have occurred. We know that rickets is controlled by two masters—one dietetic, and the other environmental. In cod-liver oil, we have a specific both for the prevention and cure of rickets. The origin of its use is obscure, but we know that its anti-rachitic properties were appreciated early in the eighteenth century. Mellanby placed the use of cod-liver oil on an experimental and proven basis. The work in this country, particularly by Parks, has shown conclusively that sunlight or the artificial mercury vapor quartz rays equally prevent and cure rickets. When one discusses, however, the fundamental question as to what prevents the deposition of lime salt in the bones, we have not arrived at a final answer. On the other hand, we have various studies on salt and inorganic salt metabolism in relation to bony development, and have accumulated a great deal of evidence.

During the past three years, Doctor Martha Jones of the University of California Medical School has been carrying on a series of experiments with puppies which has conclusively shown that the diet of the mother during pregnancy has a very marked effect on the tendency of the puppies to develop rickets. In our experience, brood bitches which are fed on a mixed food containing a liberal amount of inorganic salts give birth to normal puppies, while the same animals fed on a diet low in calcium, produce litters of puppies which have a marked rachitic tendency. These puppies frequently develop severe rickets on what appears to be a well-constituted diet. It is a well-known fact that infants, as well as animals, vary tremendously in their susceptibility to rickets. Some infants will develop rickets to a marked degree on the same diet and under identical conditions on which others develop normally. The same is true of experimental puppies. In such cases the fault unquestionably lies in the individual and not in the diet or environment. Metabolic studies conducted in our laboratory, as well as in those reported by other investigators, showed that the distribution of calcium and phosphorus between the urine and feces in a rachitic child or puppy is different from that which occurs in the normal individual—a much larger proportion of both elements being excreted in the feces of the former than in those of the latter. In rickets the alkalinity of the feces and urine is increased with a marked decrease in the retention of calcium salts. Experimentally, this condition, which ultimately results in rickets, can be produced in a puppy on a diet which appears to be adequate, in respect to protein, fat, carbohydrates, vitamins, and inorganic salts, but contains an alkaline salt mixture, and may be cured with no change in diet or environment other than the addition of sufficient HCl to neutralize the excess of alkali. Here, again, individuals of the same litter vary tremendously in their reaction to acids and alkalis. In certain individuals who develop rickets quickly and to a marked degree, very large doses of acid are required to reduce the h ion concentration of urine and feces to neutrality. On the other hand, other individuals can tolerate relatively large quantities of alkali and not excrete alkaline urine and feces. In rickets there appears to be a very nice adjustment between the acids and bases and an individual's ability to store bone-forming elements. If rickets can result from too high a degree of alkalinity in the intestinal tract, it is conceivable that this condition may be the result of an insufficiency of HCl in the gastric secretion. Those infants who have relatively little HCl may develop normally when breast-fed, but if the diet is changed to foods having a higher buffer content and potential alkalinity, such as cow's milk, the amount of acid present may be insufficient for normal mineral metabolism. Faber has shown that between 50 and 60 cc. of N/10 HCl are required to reduce the H ion concentration of a given quantity of cow's milk to a pH of 5.0, while 15 to 20 cc. of N/10 acid are sufficient to reduce the pH of a like quantity of human milk to the same value. If rickets is due to faulty absorption of the calcium salts in the intestines as the result of too high a degree of alkalinity, we can explain why artificially fed infants are more prone to the disease than those who are breast-fed. Individual difference in HCl secretion may also explain why one of a pair of breast-fed twins is rachitic and the other not.

While diet and hygiene are important factors in the production and cure of rickets, I am of the opinion that the diet of the mother during pregnancy and lactation is equally important in determining the susceptibility of an individual. Statistics show that the average American dietary is low in minerals, especially calcium. This is

particularly true of the poorer classes in large cities who cannot afford sufficient quantities of the mineral-rich foods, such as fresh fruit, green vegetables, and milk. In the light of our experimental observations which agree well with clinical experience, we dare to believe that when the dietaries of expectant mothers are planned to include liberal quantities of inorganic constituents in their proper proportions, a long step will have been taken toward the eradication of this disease which has baffled the medical profession for so many generations. If this theory is proven true, rickets of the future will be the responsibility of the obstetrician rather than the pediatrician.

ROLLA G. KARSHNER, M. D. (1136 West Sixth Street, Los Angeles)—As Doctor Chappel has justly stated, the x-ray is distinctly valuable in the recognition of rickets. However, a number of conditions must be excluded. It occurs most commonly at an age when other bone dystrophies, likewise manifesting multiple and symmetrical lesions, make their appearance. The differentiation is made by directing attention to the epiphyseal line. In rickets this is profoundly disturbed. It becomes softened and spreads, often with a roughly saw-tooth appearance. The end of the diaphysis resembles an inverted saucer, most marked in those joints where the mechanical forces are in line with the shafts. The epiphysis itself is not disturbed except possibly as a result of generalized atrophy. Often the lime salts are slightly condensed at the epiphyseal line. There is swelling of the periarticular soft tissues. The above changes cause limitation of motion and a general atrophy of the bones with softening, bending, and multiple fractures often resulting from non-use and impaired nutrition. Rarely there is periostitis in the acute stage. The chest will show the rosary even when it is not demonstrable clinically, and in severe cases one may see atalectatic strips in the lung beneath the costochondral junctions. With proper treatment, calcium is deposited in the periosteum and the cartilagenous epiphyses, in the latter case not as a direct continuation of the bone, but somewhat separated so that the picture may be confused with scurvy. In healed cases one often notes thickening of the cortex; for example, notably along the inner border of the shaft of the tibia and transverse lines of density in the ends of the shafts. The epiphyses remain expanded.

In chondrodystrophy the bones are shortened, dense; fractures are rare. The ridges for muscular attachments are enlarged. The joints are normal. The hands, feet, and changes in the base of the skull are unique. The shafts of the bones expand abruptly at the epiphyses, and ossification is deficient, irregular, often with localized overgrowths. The changes in cretinism are somewhat similar to those of chondrodystrophy.

Hereditary deforming chondrodysplasia (multiple oxostoses) often involves the epiphyses much as chondrydystrophy, the differential point being the formation of tumors of various density arising from the cortex, most frequently juxtaepiphyseal, and pointing away from the epiphysis. Osteopsathyrosis presents markedly diminished bone density, deformities, fractures, but no changes in the joints or epiphyses. In congenital lues in infants the destruction is in the epiphyses, principally, however, sharply circumscribed on the diaphyseal side at the junction of the periosteum and epiphyseal cartilage. There is usually periostitis, but no saucer-like expansion of the epiphyses. Scurvy presents intact joints and epiphyses with the destructive Trümmer zone simulating a second epiphyseal line, and there are frequently subperiosteal hemorrhages. Nutritional disturbances and arthritis (Still's disease) do not involve the epiphyseal lines.

WILLIAM SIDNEY BOWERS, M.D. (1136 West Sixth Street, Los Angeles)—In recent years preventive medicine, particularly in pediatrics, is rapidly advancing. I am very glad to see Doctor Chappel stress the point of an early diagnosis and treatment in rickets before marked deformity has occurred. In the more advanced cases, the parents frequently diagnose this disease, but then such marked deformity has occurred that, although proper curative and orthopedic treatment is instituted, it is almost impossible to overcome the deformity.

I do not agree that we have not advanced in our knowledge of the etiology of rickets, for in the past five years we have acquired considerable valuable data concerning this disease. The outstanding factors in the etiology are both dietetic and environmental. The determining factors in the diet are not entirely settled, but depend upon certain salt combinations—hydrogen ion concentration, and the presence or absence of an unknown constituent closely associated with fat soluble A vitamin. The factor of environment involved is the presence or absence of radiant energy.

The seemingly well child mentioned as developing rickets on a well-balanced diet, with sunshine, fresh air, and exercise, would not have developed it, I feel sure, had cod-liver oil, or even radiant energy in some form, been properly administered, or possibly egg-yolk. Now, while this preventive treatment need not be instituted with all infants, certainly the pediatrists and others practicing with infants should recognize certain conditions which predispose to rickets, namely, prematurity, pigmented skin, and extra-rapid growth. In these cases proper preventive treatment against rickets should be instituted.

Regarding the early recognition of rickets, I believe it can be diagnosed clinically in the majority of cases as early, and in some cases earlier, than with the roentgenogram. In other words, a negative roentgenogram does not rule out early rickets, and while the roentgenogram is a valuable aid in the diagnosis in many cases, it is not a necessity in all cases. The blood inorganic phosphate findings are quite characteristic in rickets, but not pathognomonic. Many recent investigators have emphasized the difficulty of establishing a diagnosis of rickets in the early stages from the clinical findings, but certainly if the findings warrant a probable diagnosis of rickets, it is better in this disease to err on the safe side and treat the case for rickets, for this treatment in itself is harmless.

The early signs of rickets are cranio-tabes, differentiating this from congenital ossification; rachitic rosary, ruling out scorbutic rosary, and cranial bossing, ruling out luetic bossing. I believe such findings as flabby musculature, wide fontanel with surrounding softening, and delayed dentition, when occurring in conjunction with some of the above mentioned signs, are of aid in the diagnosis, but existing alone are of little value. Enlarged epiphyses, Harrison's groove, bent limbs, "pot-belly," delayed closing of fontanel, occur relatively late in the disease. Headsweating, anemia, and palpable spleen are not dependable findings.

The orthopedic treatment as outlined by Chappel would seem conservative, and I would expect it to bring results.

H. H. MARKEL, M.D. (380 Post Street, San Francisco)—Doctor Chappel's paper is of peculiar interest to me, for I have recently had a family of three children under my care for knock-knee. The first child, a boy of 8, was so far advanced that an osteotomy of the femora was necessary. The second, a girl, needed casts only, which were later wedged on the outer side, and then wore braces. The youngest, a girl of 3, is fat and rosy and apparently well, but if she continues to be fed as the other children were she will develop a knock-knee. But I have given her mother a shotgun prescription of cod-liver oil, fresh meat and vegetables, and *sunshine,* and I expect to prevent her deformity from appearing. Another thing I have noticed is that all children who come to me for bow-legs or *knock-knees,* have *not had fresh meat to eat.* Even with a full set of teeth, the mothers all seem to fear giving them meat—why I cannot tell, except possibly a fear of choking. Recently, too, I have had consultations with Dr. Martha Jones regarding these early rachitic cases. She has assured me that her experiments have shown that deficiency of calcium metabolism in the mother in the late stages of pregnancy is responsible for rachitic changes in the young. This is manifest in the well-known saying, "For every child a tooth." I am sure that there are many more cases of rickets in California than we have formerly considered, even though we have an abundance of sunshine and fresh fruits during nearly the whole year—but we probably do not make the best possible use of them.

Physician (to rich patient)—You're all run down. I suggest that you lay off golf for a while and get a good rest at your office.—Life.